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Significant growth of adrenal lymphangioma: A case report and review of the literature

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ABSTRACT

INTRODUCTION: Adrenal lymphangiomas are benign malformations of lymphatic vessels of adrenal gland. Adrenal lymphangiomas are very rare, although lymphangiomas are most commonly located in the neck, axillary region and mediastinum.**PRESENTATION OF CASE:** A 44-year-old woman presented to the outpatient clinic with dizziness, headache during the last 2 year. We report a significant growth case of an adrenal lymphangioma removed by transperitoneal laparoscopy. At laparoscopy, a well-margined multicystic lesion was found at suprarenal area with nonviscous, brown colored fluid. The cystic mass was measured as 5.5 × 3.0 cm and histopathological diagnosis was cystic lymphangioma in the right adrenal gland. On immunohistochemical examination, D2-40 cytoplasmic staining was positive, whereas calretinin and CD34 were negative, thus, confirming their lymphatic nature. At 11 months of follow up, the patient was recurrence free.**DISCUSSION:** Lymphangiomas are benign malformations of lymphatic vessels and subtype of endothelial adrenal cysts. Adrenal cysts are histologically classified into four main groups: endothelial cysts (45%), pseudocysts (39%), epithelial cysts (9%), and parasitic cysts (7%). Endothelial cysts are divided into two subgroups: lymphangiomatous and angiomatous cysts. Lymphangiomatous adrenal cysts are also known as adrenal lymphangioma. Management of larger lesions or lesions causing symptoms may require surgical resection to determine diagnosis or relieve symptoms.**CONCLUSION:** Lymphangiomas are most commonly located in the neck, axillary region and mediastinum, which are rare at adrenal gland. Transperitoneal laparoscopic removal of the adrenal lymphangiomas is regarded as a safe, effective and minimally invasive approach. In our case, laparoscopic removal of the tumor was successfully performed via a transperitoneal approach.© 2015 The Authors. Published by Elsevier Ltd. on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Adrenal lymphangiomas are non-functioning benign malformations of lymphatic vessels that originate from an abnormal embryologic development of the lymphatic system, which are rare and usually asymptomatic. Its incidence in autopsy series varies between 0.064 and 0.18% but 6 case in Korea [1]. Lymphangiomas are most commonly located in the neck, axillary region and mediastinum (95%). The remaining 5% are found in the abdominal cavity [2]. As imaging techniques have improved, adrenal lymphangiomas usually appear as incidental findings at abdominal ultrasonography and computed tomography scan examinations. The most effective treatment option is surgical removal of the tumor. Small asymptomatic nonfunctioning cysts should be treated conservatively. We

present a very rare significant growth case of an adrenal lymphangioma that was removed by transperitoneal laparoscopy.

2. Case report

A 44-year-old woman presented to the outpatient clinic with dizziness, headache during the last 2 year. On examination, the patient showed no abnormality. The patient with adrenal masses were evaluated and screened for hyperaldosteronism, pheochromocytoma, and hypercortisolism with plasma renin activity/plasma aldosterone concentration, plasma normetanephrine and metanephrines, and urinary free cortisol. Laboratory data showed no appreciable abnormality of biochemical test. A computed tomography (CT) scan revealed a 3.0 × 2.7-cm hypodense non-enhancing lesion on the upper pole of the left kidney, which was suspected to be an adrenal cyst (Fig. 1A). And then 1 year later, we repeated CT scan. CT scan revealed a 6.0 × 2.7-cm more larger hypodense non-enhancing lesion on the upper pole of the left kidney (Fig. 1B).

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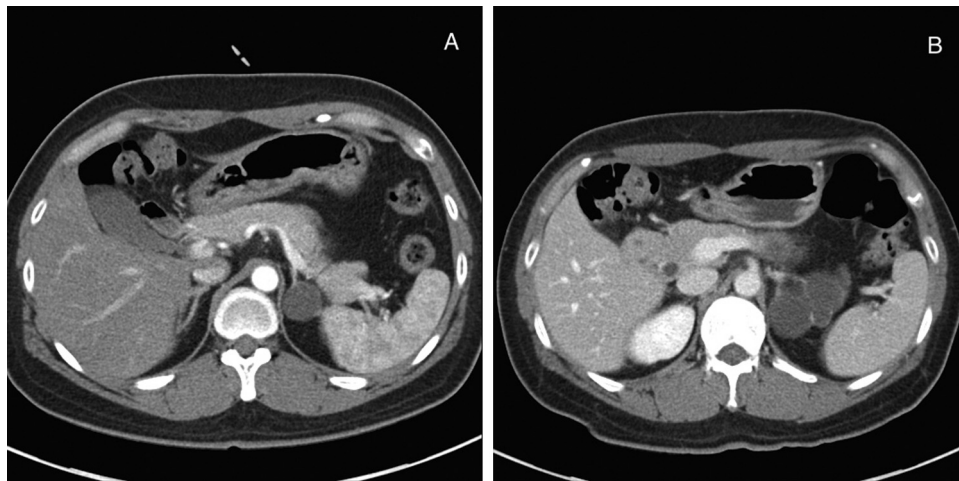


Fig. 1. Contrast-enhanced computed tomography (CT) scan shows (A) a low density mass of 3.0×2.7 cm in diameter in the left adrenal gland and (B) hypodense, lobulated, increasing mass of 6.0×2.7 cm in diameter 1 year later.

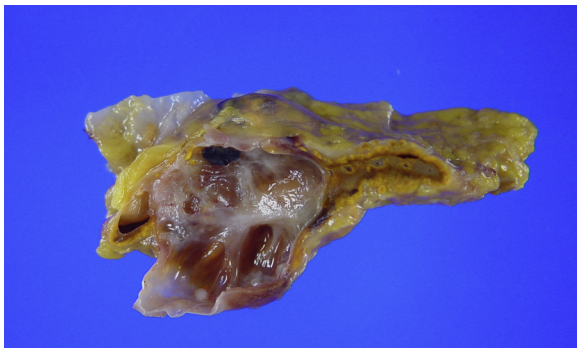


Fig. 2. Macroscopic appearance of a adrenal lymphangioma. Photograph reveals a multi-septae and smooth cystic lesion without solid portion in adrenal gland.

Laparoscopic removal of the tumor was successfully performed via a transperitoneal approach. For the first trocar insertion, a 1.5-cm transverse incision was made at the periumbilical area. And pneumoperitoneum was maintained at 12–14 mmHg with carbon dioxide (CO_2). Two additional trocars were subsequently inserted in the subcostal position between the mid-clavicular line and the anterior axillary line. Following confirmation of the location of the tumor, the tumor was successfully mobilized and removed using an ultrasonically activated scalpel and clip. Total adrenalectomy

was done. The specimen was placed in a custom made bag and removed through one of the port incision sites. A drainage tube was placed for 2 days. No complications were observed either intra- or post-operatively. On pathologic examination the adrenalectomy specimen measured $5.5 \times 3.0 \times 2.5$ cm. The cyst was filled with clear, nonviscous, brown colored fluid (Fig. 2). Microscopic examination revealed a multi-cystic lesion laid with flat endothelial cells adjacent to the normal-appearing adrenal cortex. The cystic spaces occasionally contained proteinaceous material, lacking red blood cell content, and the cyst wall contained adrenal cortex (Fig. 3A). On immunohistochemical stains, D2-40 cytoplasmic staining was positive, whereas calretinin and CD34 were negative, thus, confirming their lymphatic nature (Fig. 3B). Histopathological diagnosis was cystic lymphangioma in the right adrenal gland. No further evidence of lymphoproliferative process was detected on follow-up.

3. Discussion

Lymphangiomas are benign malformations of lymphatic vessels and subtype of endothelial adrenal cysts. Adrenal cysts are histologically classified into four main groups: endothelial cysts (45%), pseudocysts (39%), epithelial cysts (9%), and parasitic cysts (7%). Endothelial cysts are divided into two subgroups: lymphangiomatous and angiomatous cysts. Lymphangiomatous adrenal cysts are also known as adrenal lymphangioma [1]. Lymphangiomas of the

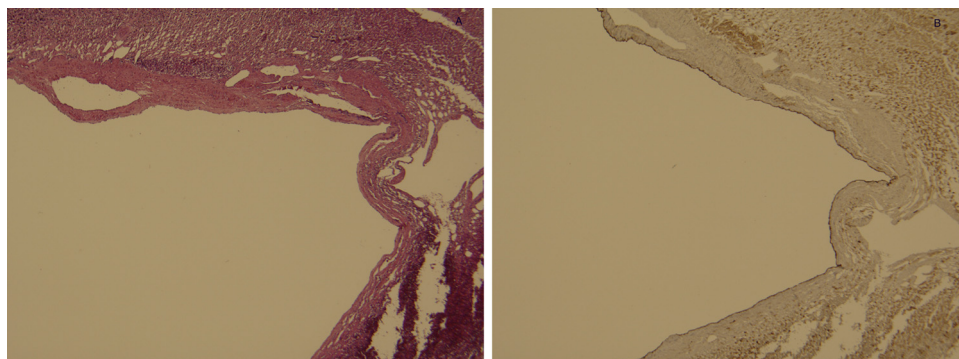


Fig. 3. (A) Histologically cystic lesion covered by thin, flat endothelial cells (H&E stain, $\times 200$). (B) Flat endothelial cells are positive for D2-40, a marker of lymphatic endothelium.

adrenal gland are rare. They are characterized by multiloculated cystic- and endothelial-lined cavities.

The etiology and pathogenesis of adrenal lymphangioma are still unknown and matters of debate. The most favored theories are: malformation of lymphatic channels, ectasia of lymphatic channels, obstruction of proximal lymphatic channels, and a cystic degeneration in a hamartoma [3].

Lymphangiomas can occur at all ages, with the peak incidence between the third and sixth decades of life, as seen on our group of cases.

Adrenal lymphangiomas are usually asymptomatic. If they are symptomatic, symptoms are usually related to size and position of adrenal lymphangiomas, and can include abdominal pain, gastrointestinal disturbance, palpable mass, and headache [4].

Histologically, all adrenal lymphangiomas showed a typical multicystic architecture composed of irregular dilated spaces lined by flattened, bland simple endothelial cells. But histologic characterization is not enough for definite diagnosis of adrenal lymphangioma, so immunohistochemical examination has to be made to prove the lymphatic origin of the cyst. D2-40 is a monoclonal antibody to the transmembrane mucoprotein which is expressed by lymphatic endothelial cells among others. D2-40 shows immunoreactivity to only lymphatic endothelium, so it is a specific marker for lymphatic origin [5].

On ultrasound, adrenal lymphangioma is a well-marginated, anechoic, cystic lesion typically located on the suprarenal lesion, whereas computed tomography demonstrates as non-enhancing, low-density (0–20HU) lesion with smooth borders and thin wall. On magnetic resonance imaging, uncomplicated adrenal cysts are low in signal intensity on T1-weighted images and high on T2-weighted images, but more complicated cysts show high signal intensity on both T1- and T2-weighted images [1,6]. In our case, during 2 years, headache and dizziness persisted and there was no specific abnormality on examination so the patient was taken for CT scan. CT was accurate in characterizing all lesions as having cystic components, and demonstrated that the cyst was originated from adrenal gland rather than kidney.

Some authors recommend aspiration of the contents of adrenal cysts for their diagnosis and management instead of surgical excision if the suspicion of malignancy is low, or the lesion is nonfunctional and asymptomatic [7]. If the appearance remains unchanged after 18 months without evidence of a clinically or biologically active tumor, the conservative management is justified [8]. But, management of larger lesions or lesions causing symptoms may require surgical resection to determine diagnosis or relieve symptoms. On cross-sectional imaging, adrenal cortical carcinomas tend to be larger than benign adrenal tumors with an average size of 10–12 cm on presentation. Indeed over 90% of ACCs are greater than 5 cm. In incidentally detected adrenal tumors, size is a relative indicator of malignancy, with 4%–5% of tumors less than 4 cm, 10% of tumors larger than 4 cm, and 25% of tumors greater than 6 cm found to be adrenal carcinomas. Given the relationship between adrenal tumor size and malignancy, it is currently recommended that adrenal tumors greater than 4–6 cm be surgically excised [9]. Therefore, we can not completely rule out malignancy because of tumors larger than 6 cm and 3 cm increase in a year. So an adrenal cyst was surgi-

cally removed. Transperitoneal laparoscopic removal of the adrenal lymphangiomas is regarded as a safe, effective and minimally invasive approach. In our case, laparoscopic removal of the tumor was successfully performed via a transperitoneal approach. No complications were observed either intra- or post-operatively. No further evidence of lymphoproliferative process was detected on follow-up.

Conflicts of interest

No potential conflict of interest relevant to this article was reported.

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Ethical approval

This paper is a case report. No ethical approval needed.

Consent

We have written informed consent from the patients.

Author contribution

Jun Min Bae contributed reports retrieval and drafting of this manuscript. Kiho kim contributed surgical procedures of this case report. Jong Im Lee contributed pathological analysis.

Guarantor

Ki Ho Kim.

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